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Rowell syndrome in pregnancy: A diagnostic challenge

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Abstract
Rowell syndrome is comprised of erythema multiforme-like lesions in association with lupus erythematosus along with serologies of speckled antinuclear antibodies, positive rheumatoid factor, positive anti-La/ anti-Ro and the clinical finding of chilblains.[1] Rowell syndrome is currently classified as a subtype of chronic cutaneous lupus erythematosus.[2] The majority of cases are seen in women aged 20-70 years. The ratio of affected women to men is 8:1. Indian patients have a median age of 23 compared to age 32 worldwide and are less likely to be discoid lupus.[3]
We report a case of Rowell syndrome in pregnancy. A 29 year old primigravida at the period of gestation 15 weeks developed vasculitic lesions, erythema multiforme-like lesions in palms, soles, oral ulceration suggestive of LE lesions. She also had Alopecia universalis, Taenia corporaris and scabies masking the lesions of Rowell syndrome which was a diagnostic challenge. Laboratory investigations revealed ANA+1:640 speckled, Anti Ro+++, Anti La +++. The histology of the targetoid lesions was consistent with erythema multiforme. The patient was successfully managed on low-dose aspirin and hydroxychloroquine. Lesions showed significant improvement in one month. Early diagnosis, multidisciplinary team approach, and intensive monitoring during the antenatal period were key to the successful outcome of patient who had normal vaginal delivery of female child at term.
This case reflects that we should suspect Rowell syndrome even in young patients without precipitating features and further cases must be reported to help increase knowledge of the pathophysiology of Rowell syndrome.

Keywords: Erythema multiforme, lupus erthematosis, targetoid lesions, ANA +, Anti Ro+

Introduction
In 1963 Rowell *et al*. described a syndrome characterized by lupus erythematosus (LE), erythema multiforme (EM)-like lesions, positive tests for rheumatoid factor (RF) and speckled antinuclear antibody (ANA).
The majority of cases are seen in women aged 20-70 years. The ratio of affected women to men is 8:1. Indian patients have a median age 23 compared to age 32 worldwide and are less likely to be discoid lupus [3]. The major criteria include LE (systemic, discoid or subacute), EM (with or without mucosal involvement), Speckled pattern of ANA and the minor criteria include Chilblains, Anti Ro/SSAor anti-La/SSB, Positive rheumatoid factor.
The higher incidence in females has been attributed to oestrogen signaling, the composition of the microbiome, and increased levels of toll-like receptor TLR7 in immune cells [5-6].

Case report
A 29-year-old women presented to antenatal OPD with amenorrhea since 4 months and a history of dusky red lesions mildly itchy, scaly rash on palms, soles, abdomen and upper arms for 2 months. She also developed fluid-filled lesions on these sites and painful oral ulcers.
The patient had a history of itching in the whole body for 10 days which was more during the night. She also had a history of painful swelling on fingertips which aggravated in cold weather (chilblains)
There was no history of photosensitivity, joint pain and malar rash.
On general physical examination patient had pallor and alopecia universalis (was wearing a wig) Systemic examination was normal. On Dermatological examination, there were red raised patches with scaling of the overlying skin and vasculitic lesions on palms and arms. There were erythematous discrete macules, papules over hands and legs which were targetoid in appearance. Mucocutaneous examination revealed erythematous painful erosions on buccal mucosa, tongue and palate.

The patient also had scabies (tiny vesicles over the web of the finger with itching) and Tinea (ring shaped red scaly patches with indistinct edges on face, abdomen and groin area).
The patient had multiple dermatological manifestations which made it difficult to conclude a diagnosis. Laboratory investigations revealed: Blood group B positive, HIV-NR, HBsAg-NR, Anti HCV-NR, VDRL-NR,S. TSH-1.3uIU/ml ,OGCT: 98 mg/dl, CHG, LFT,RFT ,24 hr urine protein were done and they were normal. Leukocytes :7.1 * 10^3, Hemoglobin: 9.3g/dl, ANA: positive (+++) Speckled, Anti-ds DNA: Negative, Anti-RNP/SM: Negative, Anti-Smith: Negative, Anit-SS-A/Ro: Positive (+++) strongly positive, Anti-SS-B/La: +++(strongly positive), Anti-nucleosome: Negative, Anti-histone: Negative, Complement C3 *:149 (84-168), Complement C4 *:26 (15-50). Rheumatoid factor: Negative. Histopathological examination revealed targetoid lesions. The patient was managed by starting her on Tablet Hydroxychloroquine 200 mg HS and low dose aspirin (75 mg OD). She was given Tab Dexchlorpheniramine 6 mg OD SOS, 5% permethrin lotion, 1% Ivermectin soap, Luliconazole 1% lotion BD for 6 weeks, Ciclopirox alamine cream for LA BD, Iron folic acid prophylaxis and Calcium supplementation.
The follow-up included maternal and fetal.

Maternal follow-up included: Hydroxychloroquine was continued throughout the pregnancy and there was a significant improvement in lesions, BP was normal throughout the pregnancy (ranging from 120-124/72-80), Proteinuria was not detected in urine (24 hr urine protein was done twice in pregnancy).

Fetal follow-up included: CMF scan was done around 20 weeks and was normal, Quadruple screen was negative, Fetal echo was normal, Growth scans were done around 32 weeks and 36 weeks which corresponded to the period of gestation. As patient was started on Hydroxychloroquine regular ocular examination was done as this drug can cause visual loss due to retinal damage. No significant abnormality in fetus (heart block) was noted. The patient reported at 38 weeks with spontaneous onset of labor pains. The labor was managed partographically.
She delivered a Female child, Birth weight 2.8 kg with Apgar 8 at 1 min and Apgar 9 at 5min. There was no gross congenital anomalies and placenta and cord were normal. There were no intrapartum and postpartum complications. The patient was discharged at post-natal day 2 and was continued on Hydroxychloroquine and was advised follow up after 2 months. The baby was also examined for any conduction defects(heart).

Discussion

Rowell syndrome is a rare entity in which patients develop LE and EM like lesions and a specific immunological pattern. In our case report the patient was booked early in pregnancy and was diagnosed early with Rowell syndrome. She was started on Hydroxychloroquine timely which resulted in successful maternofetal outcome of our patient as Rowell syndrome is known to flare up in pregnancy.
In literature there are very few cases of Rowell's syndrome which are reported. In most of cases due to lack of awareness of this syndrome most of the patients go undiagnosed and hence develop multiple complications in later half of pregnancy. Both mother and baby are at high risk of adverse pregnancy outcomes including preeclampsia, preterm delivery, pregnancy loss and intrauterine growth restriction. The predictors of adverse pregnancy outcomes include active maternal disease, nephritis, proteinuria, hypertension and thrombocytopenia [7]. Some cases have also reported neonatal lupus, in mother affected with Rowell syndrome. Preeclampsia affects 16% to 30% of SLE pregnancy compared with 5% to 7% in healthy women. The high risk of preeclampsia in SLE pregnancy is compounded by the difficulty in differentiating it from lupus nephritis [8].

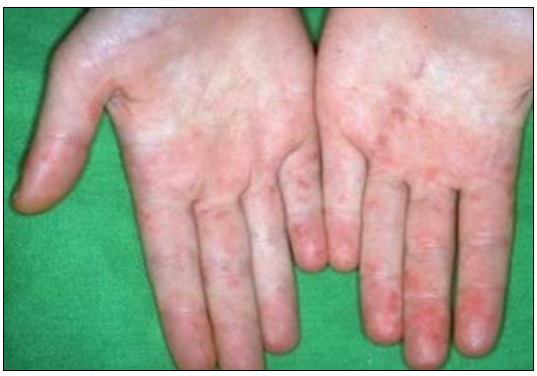

Fig 1: Vasculitic LE lesions

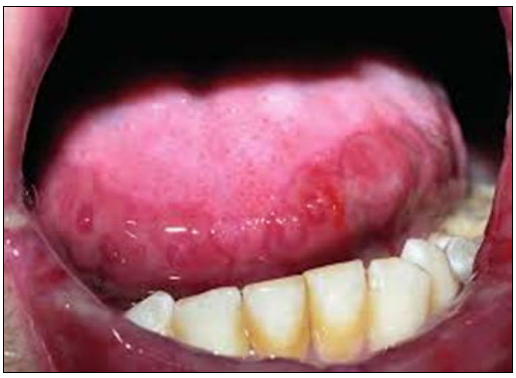

Fig 2: Oral ulcer EM lesions

Conclusion

We know that pregnancy may trigger an exacerbation of SLE and cutaneous lesions of different severity may be seen. Rowell syndrome is rare, as more cases will be reported the existence of this entity will be further clarified. Despite the refined diagnostic criteria, recent literature has debated on whether RS is an overlap syndrome, a real association, or coincidence of DLE and EM [9].
This case reflects that we should suspect Rowell syndrome even in young patients without precipitating features and further cases must be reported to help increase knowledge of pathophysiology of Rowell syndrome.

Author's Contribution: Not available

Conflict of Interest
Not available

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